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# Lung function from school age to adulthood in primary ciliary dyskinesia

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Shareable abstract (@ERSpublications)

Lung function in children with PCD is reduced by the age of 6 years and further declines during the growth period. It is essential to develop strategies to improve prognosis. <https://bit.ly/34EBekm>

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## Abstract

Primary ciliary dyskinesia (PCD) presents with symptoms early in life and the disease course may be progressive, but longitudinal data on lung function are scarce. This multinational cohort study describes lung function trajectories in children, adolescents and young adults with PCD. We analysed data from 486 patients with repeated lung function measurements obtained between the age of 6 and 24 years from the International PCD Cohort and calculated z-scores for forced expiratory volume in 1 s (FEV<sub>1</sub>), forced vital capacity (FVC) and FEV<sub>1</sub>/FVC ratio using the Global Lung Function Initiative 2012 references. We described baseline lung function and change of lung function over time and described their associations with possible determinants in mixed-effects linear regression models. Overall, FEV<sub>1</sub>, FVC and FEV<sub>1</sub>/FVC z-scores declined over time (average crude annual FEV<sub>1</sub> decline was -0.07 z-scores), but not at the same rate for all patients. FEV<sub>1</sub> z-scores improved over time in 21% of patients, remained stable in 40% and declined in 39%. Low body mass index was associated with poor baseline lung function and with further decline. Results differed by country and ultrastructural defect, but we found no evidence of differences by sex, calendar year of diagnosis, age at diagnosis, diagnostic certainty or laterality defect. Our study shows that on average lung function in PCD declines throughout the entire period of lung growth, from childhood

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to young adult age, even among patients treated in specialised centres. It is essential to develop strategies to reverse this tendency and improve prognosis.